Primary Mucinous Cystadenoma Arising from Behind the Posterior Peritoneum of the Descending Colon in a Child: A Case Report

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This is the first report of a primary mucinous cystadenoma (MCA) arising from behind the posterior peritoneum of the descending colon in a paediatric patient. A large intra-abdominal cystic lesion was found incidentally during renal ultrasonography in a 14-year-old girl. Imaging studies showed a 13 × 9 × 15 cm homogeneous cystic lesion with mild contrast enhancement of the wall. The cyst appeared to originate from the retroperitoneum, but was separated from the left kidney, ovary, and pancreas. At laparotomy, there was a cyst behind the posterior peritoneum of the descending colon. The cyst was successfully excised, and histopathology showed MCA. Although primary MCA in the retroperitoneum is extremely rare in children, it should be considered in the differential diagnosis of an intra-abdominal cyst, since it needs to be excised to eliminate the risk of infection, recurrence, and malignancy. [*Asian J Surg* 2003;26(4):237–9]

Introduction

Mucinous cystadenoma (MCA) is rare and has been described in the ovary, pancreas, and appendix.[1,2] Primary MCA in the retroperitoneum is extremely rare and only a few cases have been reported in adults.[2,3] To the best of our knowledge, this is the first report of primary MCA arising from the retroperitoneum in a paediatric patient.

Case report

A 14-year-old girl was found to have proteinuria during a routine school screening programme and was referred for further investigation at a local hospital. She had a blood test to check her renal function and underwent abdominal ultrasonography. Although her renal function and kidneys proved to be normal, and proteinuria was not detected again, a large intra-abdominal cystic lesion measuring 13 × 9 × 15 cm was found in the left upper quadrant during ultrasonography and she was referred to our hospital for further assessment of this lesion. On presentation, she was asymptomatic and her abdomen was non-tender. However, no mass could be palpated because she was obese. Computed tomography and magnetic resonance imaging confirmed that there was a 13 × 9 × 15 cm homogeneous cystic lesion, with mild contrast enhancement of the cyst wall, located in the retroperitoneum in the left upper quadrant of the abdomen (Figure 1). It was separated from the left kidney, ovary, and pancreas. The density reading of the fluid filling the cyst was consistent with that of water. Barium enema showed that the descending colon was displaced anteromedially, and there was no duplication. Blood biochemistry was normal without any signs of inflammation. Provisional diagnosis included mesenteric cyst, cystic teratoma, retroperitoneal lymphangioma, and duplication of the colon.

On laparoscopy, the cyst was found to be located behind the posterior peritoneum of the descending colon.

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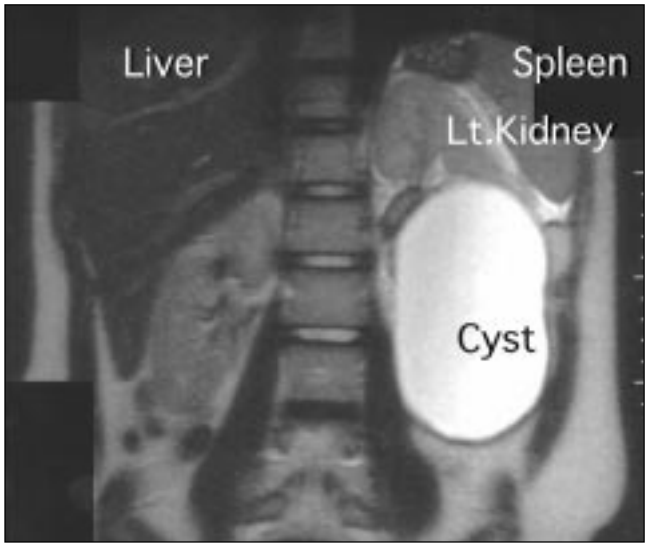

Figure 1. Magnetic resonance imaging demonstrates a large homogeneous intra-abdominal cyst.

Laparoscopic excision was attempted, but complete resection was not possible due to adhesions between the cyst and the posterior peritoneum behind the descending colon, so laparoscopy was converted to open laparotomy. The abdomen was opened through a left upper transverse incision, and full-thickness excision of most of the cyst wall was performed. Where full-thickness excision was not possible because of dense adhesions between the cyst, the peritoneum, and the colonic wall and vessels, mucosectomy was performed. The cyst could be separated easily from the kidney, peritoneum, and pancreas by blunt dissection, but tightly adhered to the posterior peritoneum behind the descending colon. The cyst did not originate from pelvic or urological structures. No other lesions were found in the retroperitoneal space or in the peritoneal cavity. The uterus, fallopian tubes, and both ovaries were normal in size, appearance, and location.

The cyst contained clear mucinous fluid with low amylase content and class I cytology. On gross inspection, the cyst wall was 3 mm to 5 mm thick and the inner surface was smooth. Histopathology showed the lining of the cyst to be a single layer of mucinous columnar epithelium (Figure 2). There was no evidence of metaplasia or dysplasia. No ectopic ovarian, intestinal, pancreatic, or other tissue was identified in the cyst. There were loose interstitial cells of similar appearance to ovarian interstitial cells in the submucosa, which stained positively for progesterone receptors (Figure 2). The final diagnosis was primary MCA behind the posterior peritoneum of the descending colon.

The patient made an uneventful postoperative recovery without additional therapy. Follow-up abdominal ultrasound 3 months and 12 months after surgery revealed no abnormalities. At the time of writing, she is 13 months' post-operative and well, without any evidence of recurrence.

Discussion

Primary MCA has been described in the ovary, pancreas, and appendix.[1,2] However, primary MCA in the mesentery or behind the posterior peritoneum of the colon is rare.[3] Its histogenesis remains unclear, but one theory postulates that it may arise from ectopic ovarian, pancreatic, or appendiceal tissue.[4] However, in our case, we could find no evidence of ectopic tissue in the cyst wall. We found mucinous columnar epithelium lining the cyst and progesterone receptor-positive, ovarian-like interstitial cells in the submucosa.[5,6] To the best of our knowledge, this is the first report of MCA arising from behind the posterior peritoneum of the colon in a paediatric patient.

There is a report of an 18-year-old primiparous woman who had a retroperitoneal MCA with Mullerian-type epithelium and mucinous differentiation. The patient died of diffuse carcinomatosis shortly after delivering a premature baby.[7] More recently, Banerjee and Gough reported a 38-year-old woman with an MCA in the mesentery of the descending colon and a 58-year-old woman with an MCA in the mesentery of the hepatic flexure.[3] In both cases, the cyst was lined with mucinous columnar epithelium. However, in the former case, there was "borderline" histology in several areas of the cyst wall, and she developed metastases to mediastinal lymph

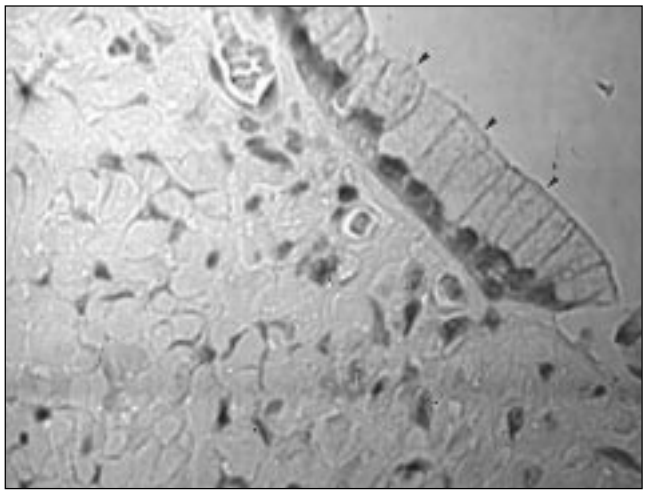

Figure 2. Benign columnar mucinous epithelium (arrowheads) lining the cyst wall and progesterone receptor-positive, ovarian-like interstitial cells (arrows) in the submucosa. (Progesterone receptor immunohistochemistry, × 200.)

nodes 4 years after diagnosis. Based on these clinical findings, MCA seems to have malignant potential. Thus, the treatment of choice for MCA is complete excision to eliminate the risk of malignancy, even though this complication is probably rare. Marsupialization, internal drainage, or simple aspiration are not recommended because of the increased incidence of infection and recurrence.[1]

The differential diagnosis of a cystic lesion in the mesentery includes a variety of pathologies. Pancreatic pseudocyst can be excluded because a pseudocyst is usually accompanied by tissue necrosis and inflammation, and does not have any epithelium. Duplication of the colon can also be excluded because of the absence of smooth muscle cells, and mesenteric cyst can be excluded because it normally originates from ectopic lymphatic tissue, and should not have any columnar cells. Retroperitoneal lymphangioma, cystic teratoma, and ovarian cyst can easily be eliminated on operative and histopathological findings.

In conclusion, although primary MCA is extremely rare in children, it should be considered in the differential diagnosis of an intra-abdominal cyst, since the cyst should be excised to eliminate the risk of infection, recurrence, and malignancy.

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